

Intrapulmonary Solitary Fibrous Tumor Masquerade Sigmoid Adenocarcinoma Metastasis

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Solitary fibrous tumor is a rare spindle cell mesenchymal tumor entity, with either benign or malignant behavior that cannot be accurately predicted by histological findings. An intrapulmonary site of origin is even rarer. We report a case of a 51-year-old woman in whom an abnormal nodule in the lower right lung was detected during staging for sigmoid adenocarcinoma. The nodule was excised and pathological examination revealed an intrapulmonary solitary fibrous tumor.

Key words: 1. Solitary fibrous tumors
2. Lung neoplasms
3. Video-assisted thoracic surgery

CASE REPORT

A 51-year-old Caucasian female was referred to our institution because of an abnormal nodule detected in her right lung. This had at first been detected four months earlier when she underwent an urgent Hartmann procedure due to refractory adenocarcinoma of the sigmoid colon (pT4N₁ stage). Due to the urgency of the surgery, the patient was not staged prior to the surgery but was initially staged after the operation. The only pathologic finding in the computed tomography (CT) of the lung was a nodule in the lower right lobe of 2.5 cm at its longest dimension (Fig. 1), and the patient was staged as cT₄N₁M₁. She received 4 cycles of chemotherapy for the sigmoid adenocarcinoma before she was referred to us for excision of what was believed to be a single metastatic nodule. Although the radiologic findings supported a benign tumor, neither positron emission tomog-

raphy-CT nor any other procedure to attempt to diagnose the nodule was performed due to the patient's willingness to undergo complete removal of the mass even if it were benign. The patient underwent video-assisted thoracoscopic (VATS) wedge excision of the tumor. She had an uneventful postoperative course and was discharged on the 4th postoperative day.

The specimen of the resected lung contained a firm intrapulmonary white-colored well-defined mass with dimensions of 2.5×2.3×2.2 cm. Microscopically, the tumor had a "patternless pattern," with proliferation of bland spindle cells in alternating hypocellular and hypercellular areas, accompanied by a collagenous stroma (Fig. 2A) with branching hemangiopericytoma-like vessels (Fig. 2B). The tumor had no evidence of increased mitotic activity (0 to 1 mitoses per 10 high power fields using an Olympus BH-2 microscope, with a 40× field, 0.5 mm diameter, and area of 0.196 mm²), any

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significant atypia, or necrosis. Immunohistochemically, the tumor cells were positive for vimentin, CD34 (Fig. 2C), and Bcl-2. A few cells were also focally and weakly positive for estrogen and progesterone receptors. The index of cellular proliferation with the antibody Ki-67 is low and estimated at <1% of the cellular population. Immunohistochemistry staining of the tumor cells was negative for alfa-smooth muscle actin (SMA), muscle specific actin (HHF-35), h-caldesmon, CD117 (c-kit), S-100 protein, epithelial antigens (wide spectrum cytokeratins [AE1/AE3] and epithelial membrane antigen), and thyroid transcription factor-1. The resection margins were free of disease. One year after the operation, the patient is well, with no evidence of recurrent disease.

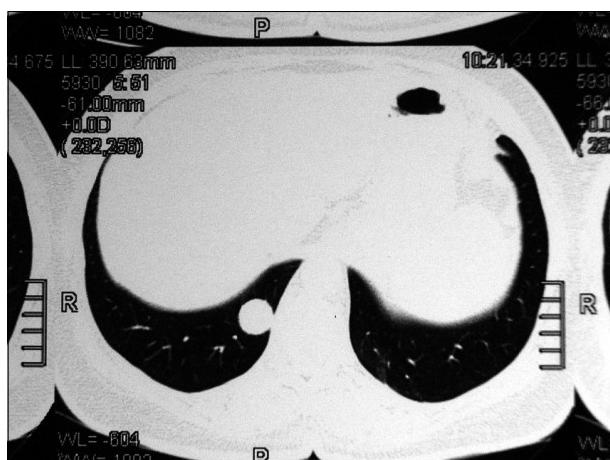


Fig. 1. Computed tomography scan showing a well-defined tumor in the lower right lobe of the lung.

DISCUSSION

Solitary fibrous tumors (SFTs) were first pathologically described by Klempner and Rabin in 1931. To the present, about 800 cases have been reported in the English medical literature [1].

SFT is the preferred term for an uncommon, but histomorphologically distinctive spindle cell neoplasm, that was identified in the past as fibrous mesothelioma, localized fibrous mesothelioma, localized fibrous tumor, localized mesothelioma, pleural fibroma, solitary fibrous mesothelioma, or submesothelial fibroma [1]. There are reports of SFTs arising either from mediastinal, diaphragmatic, or parietal pleura, or from within a lung fissure and in various extrapleural locations, such as the retroperitoneum, mediastinum, thyroid gland, nasal cavities, meninges, or parietal surfaces of the intra-abdominal viscera. The intraparenchymal or endobronchial location is a rare occurrence [1]. Less than 20 cases of intraparenchymal SFTs have been reported in the English literature [1-5]. To prove and establish the fact that these lesions are indeed of pulmonary origin, it is important to consider the clinical, radiologic, and pathologic findings to demonstrate the lack of continuity with the visceral pleura and to exclude an endophytic growth in a pleural-based lesion [1,2,4].

SFTs of the pleura and lung occur predominantly in adults. Nevertheless, there are rare cases of intrapulmonary SFTs reported in childhood [6]. They are usually found incidentally. Possible symptoms include cough, pain and dyspnea, and dig-



Fig. 2. Histologic appearance of solitary fibrous tumor. (A) The lesion is well delineated from the lung parenchyma, which is shown on the left (H&E, $\times 25$). (B) The tumor is characterized by a growth of bland spindle-shape cells, with variable cellularity and branching hemangiopericytoma-like vessels in the stroma, shown in the center (H&E, $\times 100$). (C) Strong and diffuse immunoreactivity of the tumor cells for CD34 ($\times 100$).

ital clubbing. Hypoglycemia and seizures can be encountered in some cases due to secretion of insulin-like growth factor II by the tumor [1-3,5]. Hemoptysis due to SFT as well as hypertrophic osteoarthropathy secondary to SFT of the lung has also been reported. There is no association with smoking or asbestos exposure. Due to its non-specific symptoms, it can potentially mimic solitary pulmonary metastasis, as was the case with our patient.

On gross examination, the tumor is generally well-circumscribed with bulging and whorled cut surfaces. The tumor size can range from 1.9 to 20 cm. Histologically, it is associated with variable patterns, the most common being the “patternless pattern.” The cellular areas with the spindle-shaped cells may alternate with hypocellular areas. The cytologic features consist of cellular areas composed of spindled to ovoid cells with scanty cytoplasm, occurring either solely or in groups. Immunohistochemistry plays a substantial role in supporting the diagnosis made by morphologic features and is also helpful in differentiating these tumors from mesothelioma, neurofibroma, and other spindle-cell lesions. A differential diagnosis of intrapulmonary SFT includes, among other lesions, pulmonary adenofibroma, benign neural neoplasms, leiomyoma and leiomyosarcoma, synovial sarcoma, spindle cell thymoma, spindle cell carcinoid tumor, nerve sheath tumor, fibrosarcoma, sarcomatoid carcinoma, and sarcomatoid mesothelioma. The morphologic features and immunohistochemical profile of SFT are sufficiently distinct to allow separation from such conditions in the majority of the cases. Several studies have reported positivity of tumor cells with CD34 antibody in almost 100% of cases, and with CD99 antibody in 70% of cases, whereas Bcl-2, SMA, and epithelial membrane antigen are positive in 20% to 35% of the cases. Vimentin is positive in 90% of the cases, but is considered nonspecific [7,8].

The histologic criteria for classifying the malignant variants of SFT of the lung and pleura were described by England et al. in 1989 and Vallat-Decouvelaere et al. in 1998. They established the following features suggestive of malignancy: 1) more than 4 mitoses per 10 high-power fields, 2) presence of necrosis, 3) hemorrhage, 4) hypercellularity as detected by nuclear crowding and overlapping, 5) nuclear atypia, 6) pleomorphism, 7) stromal or vascular invasion, and 8) size ex-

Table 1. Staging system for the management and follow-up of SFT

Stage	Description and % recurrence
0	Pedunculated tumor without signs of malignancy (<2% recurrence)
I	Sessile or “inverted” tumors without signs of malignancy (<8% recurrence)
II	Pedunculated tumor with histological signs of malignancy (14% recurrence)
III	Sessile or “inverted” tumor with histological signs of malignancy (63% recurrence)
IV	Multiple synchronous metastatic tumors

ceeding 10 cm.

These pathologic features are only suggestive. The absence of these characteristics does not exclude malignant behavior. On the other hand, encapsulation, pedunculated and resectability with free surgical margins are considered to be favorable prognostic factors even in histologically malignant variants. Positive margins are associated with an aggressive clinical course and high rates of local recurrence and metastasis [1].

Resection with free margins is considered to be the treatment for SFT located either in the lung or on the pleura. Wedge resection may be accomplished by VATS or standard thoracotomy, according to the anatomic position and size of the lesion and the experience of the surgeon [2,4]. Adjuvant therapy may have a place in recurrent or systemic disease, but its benefit is undefined [5]. In general, SFTs have an unpredictable course, depending on their potential for malignancy. In large series, the recurrence rate of benign SFTs is reported to be low (1.4%), while the recurrence rate of malignant variants is reported to be higher (range, 9% to 19%) [1,5].

A staging system based on pedunculated versus sessile attachment and malignant versus benign histology that predicts recurrence has been proposed by Perrot et al. [1] (Table 1).

Due to the possibility of local recurrence and/or distal metastasis after surgical removal of the primary SFT, long-term follow-up is recommended. Local recurrence detected early is amenable to reoperation and resection, with good long-term results. The long-term survival rate for both benign and malignant variants is reported to be more than 90% [1-3,6].

In conclusion, SFTs are neoplasms that usually arise from

the pleura. An intraparenchymal or endobronchial location is a rare occurrence. Resection with clear margins is curative and final diagnosis and prognosis can be defined only after surgical resection. The prognosis of patients with rare SFTs of the lung depends on the completeness of the tumor resection. Variants of SFTs of the lung that are malignant or suspected to be malignant should be managed as lung cancer with regard to the surgical resection of the tumor and follow-up strategy.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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